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IMAGING

Shrinking Lung Syndrome Caused by Systemic Lupus Erythematosus: a Case Report

Sadettin Uslu[1]

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Abstract

Shrinking lung syndrome (SLS) is a rare manifestation of systemic lupus erythematosus (SLE), characterized by dyspnea, pleuritic chest pain, diaphragmatic elevation, and decreased lung volumes. We describe a lupus patient who presented with dyspnea. In addition, she had characteristic chest imaging findings of small lung volumes, elevated left hemidiaphragm, and thinning of the diaphragmatic crura. The patient was diagnosed as SLS, and treated with azathioprine and prednisone.

Keywords Shrinking lung syndrome · Systemic lupus erythematosus · Dyspnea

Introduction

Shrinking lung syndrome (SLS) is a rare pulmonary manifestation of autoimmune rheumatic diseases. The overall SLS prevalence in systemic lupus erythematosus (SLE) was 0.5–1.1% and reported a female:male ratio of 17:1, with a median duration of SLE of 4–6.5 years before a diagnosis of SLS was made [1].

Case Presentation

A 38-year-old female presented with dyspnea and constitutional symptoms. Five years previously, she was diagnosed with SLE. On examination, she had a fever of 38.5 °C, tachypnea (21/min), and reduced breath sounds in the left lower lung zone. On laboratory, ESR (60; range, 0–20 mm/h) and CRP (45; range, 0–5 mg/L) were increased. Autoimmune serology showed a strongly positive ANA (1:640, homogeneous staining) and positive anti-dsDNA (52; range, 0–10 IU/ml). On chest X-ray, there was an elevated hemidiaphragm and loss of volume on the left lung with normal parenchyma (Fig. 1a). Computed tomography (CT) showed thinning of the diaphragmatic crura (Fig. 1b) and the absence of significant parenchymal and pleural disease. Pulmonary function tests (PFTs) confirmed a restrictive defect (FEV1: 62%, FVC: 66%) and carbon monoxide diffusion capacity (DLCO: 57%) was reduced. The SLEDAI (SLE severity disease activity index) score was 7 (mild or moderate flare). All causes of elevated hemidiaphragm including trauma, metabolic diseases, infectious diseases, direct invasion by tumor, and neurological diseases were ruled out. According to clinical manifestations and imaging studies, the diagnosis of SLS was suspected. The patient was started on azathioprine (150 mg/day) and prednisone (20 mg/day) along with the introduction of nebulized salbutamol and theophylline. A clear improvement in the patient's general condition was observed within a few days of the treatment. At the last visit, improvement was observed in PFTs (FVC: 78%) and DLCO (70%).

Discussion

SLS is a rare pulmonary manifestation of SLE characterized by progressive dyspnea, pleuritic chest pain, elevation of the diaphragm, lung volume reduction, and restrictive pattern on PFTs. Chest x-ray often shows clear lungs with diaphragmatic elevation. Occasional basal atelectasis may be present. Computer tomography (CT) chest often shows reduced lung volumes with diaphragmatic elevation and occasional basal

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✉ Sadettin Uslu
sadouslu@gmail.com

[1] Department of Rheumatology, Ömer Halisdemir University Bor Physical Medicine and Rehabilitation, Training and Research Hospital, 51200, Niğde, Turkey

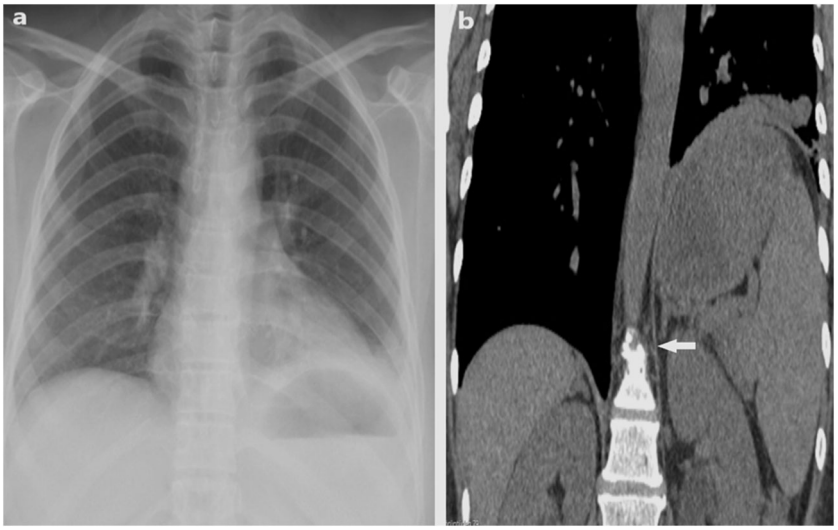

Fig. 1 **a** Chest X-ray is showing elevated left hemidiaphragm at the time of SLS diagnosis. **b** CT image is showing the thinning of the diaphragmatic crura (arrow) at the time of presentation

atelectasis but without any pleural or parenchymal abnormalities. The evaluation of diaphragm dome motion by dynamic contrast-enhanced lung MRI might be a useful in cases of diagnostic difficulty [1].

The pathophysiology of SLS is not fully understood. The immunologic mechanisms involved in other major organ involvement in SLE are not obvious in this manifestation. The underlying pathology range from loss of surfactant, diaphragmatic myopathy or myositis, phrenic neuropathy, to the most recent one of pleuritis causing reflex inhibition of deep inspiration resulting in chronic lung hypoinflation and poor pulmonary compliance [2].

No treatment has been validated, because of its low prevalence, but similar to our report, corticosteroids in combination with immunosuppressive agents (azathioprine, cyclophosphamide or rituximab) have been suggested in the case series. The rationale for rituximab use in SLS is based on the hypothesis of phrenic nerve neuropathy due to immune complex or autoantibody-mediated injury [3]. Belimumab is a human immunoglobulin G1λ monoclonal antibody which blocks the binding of soluble B lymphocyte stimulators to the B cells [4]. Belimumab might be beneficial effects, particularly in patients with severe or refractory disease, although larger cohort studies must be performed in this field to confirm the efficacy of belimumab in SLS [5]. The overall response to treatment is positive and mortality rates are low. The great majority of patients achieve clinical and spirometrical improvement after treatment [6].

In conclusion, an awareness of the existence of SLS, coupled with a high index of suspicion in lupus patients with otherwise unexplained dyspnea, or pleuritic chest pain is required to diagnose and manage this treatable condition.

Authors' Contributions Concept – S.U.; Design - S.U.; Supervision - S.U.; Resources - S.U.; Materials - S.U.; Data Collection and/or Processing - S.U.; Analysis and/or Interpretation - S.U.; Literature Search - S.U.; Writing Manuscript - S.U.; Critical Review - S.U.; Other - S.U.

Data Availability N/A

Compliance with Ethical Standards

Broad informed consent was obtained from the patient.

Conflicts of Interest The authors declare that they have no conflict of interest.

Consent to Participate N/A.

Consent for Publication N/A.

Code Availability N/A.

Peer-Review Externally peer-reviewed.

Informed Consent We received informed consent from the patient for the publication of this case.

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